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# *Clinical Proceedings*

*of the*

## CHILDREN'S HOSPITAL

WASHINGTON, D. C.

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# SHOULD VITAMIN D BE GIVEN ONLY TO INFANTS?

VITAMIN D has been so successful in preventing rickets during i fancy that there has been little emphasis on continuing its use aft the second year.

But now a careful histologic study has been made which reve a startlingly high incidence of rickets in children 2 to 14 years o Follis, Jackson, Eliot, and Park* report that postmortem examin tion of 230 children of this age group showed the total prevale of rickets to be 46.5%.

Rachitic changes were present as late as the fourteenth year, a the incidence was higher among children dying from acute dise than in those dying of chronic disease.

The authors conclude, "We doubt if slight degrees of ricke such as we found in many of our children, interfere with hea and development, but our studies as a whole afford reason to p long administration of vitamin D to the age limit of our study, fourteenth year, and especially indicate the necessity to suspect a to take the necessary measures to guard against rickets in s children."

*R. H. Follis, D. Jackson, M. M. Eliot, and E. A. Park: Prevalence of rickets in child between two and fourteen years of age, Am. J. Dis. Child. 66:1-11, July 19

MEAD'S Oleum Percomorphum With Other Fish-Liver Oils and Viosterol is a potent source of vitamins A and D, which is well taken by older children because it can be given in small dosage or capsule form. This ease of administration favors continued year-round use, including periods of illness.

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*SPECIAL REPORT*

# CHRONIC CONSTRICTIVE PERICARDITIS

## Report of the Medical and Surgical Aspects of a Case

Dr. Bernard Walsh[1]

K. H., (45-3336), a 10-year old white girl, was referred for study by Dr. Gervase Nealon of Latrobe, Pa., who had made a tentative diagnosis of chronic constrictive pericarditis.

This patient enjoyed excellent health until the spring of 1940 when she became ill rather suddenly with fever, some cough, and increasing swelling of her abdomen. The condition was considered due to pneumonia. It cleared in about a month and she seemed her usual self until the spring of 1942 when she had another episode of illness like that of 1940. There was some doubt that her abdomen ever returned to its normal state after the illness of 1942. She apparently got along well until the abdominal swelling increased to such a degree that tapping was necessary in May 1944. It was necessary to do abdominal paracentesis at approximately three-week intervals since May 1944. On each tap between 8000 and 12,000 c.c. of fluid were obtained. During the early part of 1944 she had six or seven pericardial taps with removal of 50 to 150 c.c. of non-purulent, non-hemorrhagic fluid. At the last two taps no pericardial fluid was obtained. She had no particular complaints except those referable to the considerable abdominal swelling. She had no notable joint pain or rash. There was no swelling of the feet or legs. Her appetite was at least fair.

I saw her for the first time on April 25th, 1945. She was moderately uncomfortable, apparently because of a moderate amount of ascites. There was no cyanosis. The cervical veins were prominent though the patient was semi-upright. The mucous membranes were slightly pale. The blood pressure was 90 systolic, 70 diastolic. The cardiac apex impulse could not be felt. The heart sounds were readily heard and not particularly distant in quality. There were no murmurs or friction rubs. The heart rate was regular at a rate of 120. There were a few crackles at the lung bases. The abdomen was considerably distended by fluid and satisfactory examination was not possible. There was no peripheral edema. Electrocardiogram showed auricular tachycardia with two to one block, the auricular rate being 240, and the ventricular rate 120. There

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was slight right axis deviation with somewhat low upright T waves in Lead 1, low upright T waves in Lead 2, and inverted T waves in Lead 3. The QRS complexes were not reduced in voltage.

Fluoroscopic examination with the patient standing showed a very large heart, somewhat bottle-shaped in appearance. No calcification was seen in the heart or pericardium. The amplitude of the cardiac pulsations was reduced throughout but was at least fair on the right border and best of all along the upper left border of the heart. There was no notable change in the heart size, shape, or amplitude of the pulsations

FIG. 1. K. H., X-RAY TAKEN UPON ADMISSION SHOWING LARGE CARDIAC SILHOUETTE

with shift in the position of the patient. There was no evidence of pneumonitis or pleural effusion.

The laboratory data were reported as follows: Hemoglobin 13 grams, red blood cell count 4,770,000, white blood cell count 9150. Urinalysis: Specific gravity 1.015, faint trace of albumin. Kahn and Mazzini tests negative. Total protein: 4.62 grams.

The day after her admission 7900 c.c. of straw-colored fluid were removed from the abdominal cavity. The liver was readily felt. It was hard, smooth, non-tender, with the edge 6 to 7 cm. below the costal margin in the right midclavicular line. It extended to the left midclavicular line. The venous pressure in the right arm following removal of the fluid was 220 mm.

The problem here concerned the possible causes of pericarditis with

effusion, the possible relationship of chronic pericarditis and/or myocardial disease to the ascites, and whether or not surgery would offer cure or benefit.

Pericarditis may be acute or chronic. It may or may not cause cardiac compression, and, what I think is important and very little recognized, there may be definite myocarditis associated with the pericarditis. I have come to believe on the basis of observation of several patients in the past few years that a greater or lesser degree of myocarditis occurs in most patients with pericarditis and has definite influence upon the prognosis.

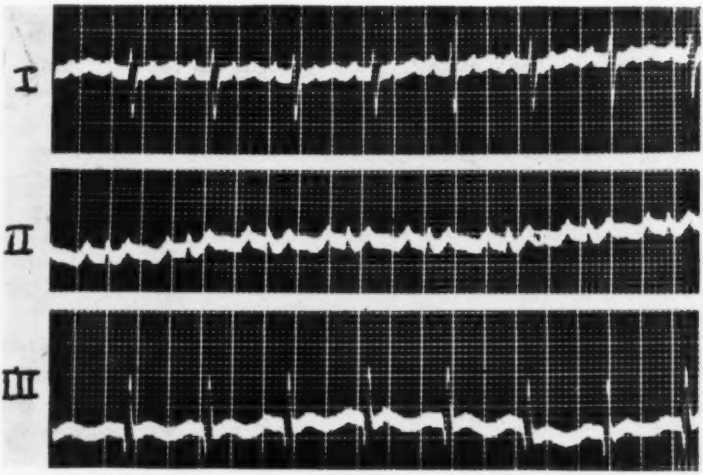


FIG. 2. K. H. (4–27–45). AURICULAR TACHYCARDIA, TWO TO ONE RHYTHM, AURICULAR RATE 240, VENTRICULAR RATE 120

Slight to moderate right axis deviation. T waves obscured by auricular waves but probably upright in Leads 1 and 2. QRS duration .08 second.

For about two years I have observed several patients who have had nonpurulent, nonspecific pericarditis whose hearts have been slightly to considerably enlarged during the time of the active pericarditis. All but two of these parients are children or adolescents. In most of them the heart has remained large following the subsidence of the acute pericarditis. They have yielded from a few drops to 100 c.c. of straw-colored fluid on pericardial tapping on one or two occasions during the course of the acute pericarditis. In others the heart size has remained unchanged. The venous pressure was at first elevated but has come to normal in all of these patients. Three or four of the total group of about 10 had evidences

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of cardiac compression and/or myocardial failure during the early period of the illness. One patient, a four year old colored child seen here two years ago, at autopsy showed a moderately thickened gelatinous pericardium, and moderate dilatation of all cardiac chambers. There were only a few c.c. of straw-colored fluid in the pericardial sac. No particular etiology was determined.

I have been impressed by the poor results from operation in several patients with definite chronic constrictive pericarditis and large hearts. No entirely satisfactory explanation for these poor results has been offered. I believe the poor results may be due to the myocarditis and cardiac dilatation, and that the chronic pericarditis may sometimes be of little functional importance.

The greatest part, I suspect nearly all, of the large cardiac shadow seen by x-ray in this patient is made up of dilated heart. I doubt very much that there is any significant amount of pericardial fluid or that the pericardium is more than a few millimeters thick. In my opinion, the pericarditis in this patient is less important than the myocarditis. There is only one way in which we can be certain how important this patient's pericarditis may be, and whether or not she may be helped by surgery, and that is by going ahead with surgical exploration. I suspect Dr. Davis will be reluctant to consider doing pericardiectomy here because of the heart enlargement. However, with the understanding by the parents that there is nothing else to offer which will result in lasting benefit and that the risk of operation is justified by the possibility of obtaining definite improvement, I have urged that Dr. Davis carry out pericardial resection.

I do not know how long the auricular tachycardia has been present, since the ventricular rate is unusually low because of the two to one block. The arrhythmia may have been present for weeks, and may possibly be a factor in the production of the cardiac enlargement and venous hypertension. Thus far quinidine has served only to slow the auricular rate from 240 to 220 per minute. The auricular tachycardia will not add to the risk of operation if the ventricular rate is maintained at 110 to 120. It is not uncommon for patients with pericarditis, particularly of the chronic constrictive form, to have cardiac arrhythmias.

*Later Note:* On May 3rd the patient was noted to have grossly irregular cardiac rhythm, rate 90 per minute. There was no particular distress, and no notable changes on examination. The electrocardiogram confirmed the impression that she had auricular fibrillation. She was digitalized. On May 6th the abdominal paracentesis was repeated and 12,000 c.c. of straw-colored fluid were removed from the abdominal cavity. The venous pressure the following day was 250 mm. of saline in both arms.

On May 9th pericardial resection was carried out by Dr. Edgar Davis. Dr. Davis will discuss the operative procedure.

The postoperative course was satisfactory. The heart rhythm returned to normal at the end of the first week. The venous pressure measurement in the arm at the time of discharge on May 20th was 190 mm. of saline. The liver edge could be felt only in the epigastrium about 3 cm. below the xiphoid process. There was little or no ascites.

The electrocardiogram May 20th showed normal rhythm, prominent P waves, and considerable right axis deviation.

By x-ray the transverse diameter of the heart was possibly slightly greater than before operation, the increase, if any, being in the region of the right auricle.

A recent note from her local physician stated that the child did well for one month, but that it was necessary to do an abdominal paracentesis on July 9th at which time 9000 c.c. of fluid were removed, and that paracentesis had to be repeated in four weeks. Her physician felt that she was definitely better. She was able to be up and around when she was not bothered by a large amount of ascites. She was eating well and her color was excellent. The heart rate was regular at a rate of 90.

*Comment:* The pre-operative opinion that this patient had chronic pericarditis and a greatly enlarged heart was confirmed. The amount of fluid in the pericardium was greater than had been estimated, but it does not seem likely that the pericardial fluid was important in the production of the symptoms and signs. It has been shown again that operation in a patient with chronic pericarditis and cardiac enlargement due to myocarditis may result in only a slight degree of benefit.

The outlook here seems poor, and it is probable that she will succumb to heart failure in the next decade. I believe that further pericardial resection would not be of benefit.

### DISCUSSION OF THE SURGICAL ASPECTS

Dr. Edgar Davis[1]

Although many patients have been greatly benefited by surgical removal of a constricting pericardium, some patients, particularly those with large hearts have not experienced complete relief of the symptoms and signs produced by the constricting pericardium. It has been my feeling that the technical difficulties of the operative procedure may have played a role in some of the unsuccessful operations, since the conventional anterior

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approach with removal of the third, fourth, and fifth cartilages together with part of the sternum has not permitted complete visualization of the anatomical structures, in particular the auricles, the superior vena cava and the pulmonary vessels. Also, in order to remove all of the pericardium safely by the usual approach it is necessary to use much traction or to remove considerable bone. Realizing the disadvantages of the usual surgical approach and after discussion with medical colleagues I ventured

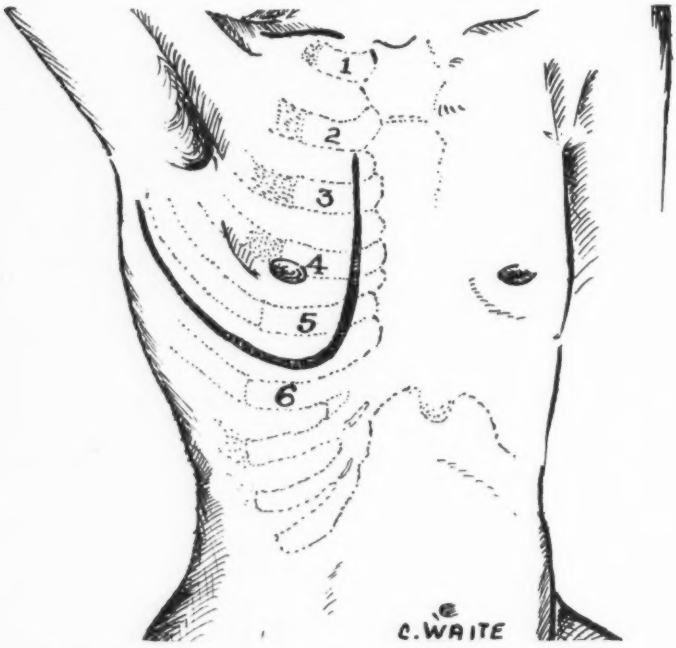


FIG. 3. K. H., ILLUSTRATING INCISION USED IN THIS CASE FOR PERICARDIECTOMY

to use a new technique in this patient, and as a result of this experience I believe this transpleural method is much better than the usual anterior approach. I believe the transpleural approach should be used, then if necessary, a similar approach could be used at a later time on the left side to complete the removal of all of the constricting pericardium. The fact that two operations rather than one might be required should not condemn the procedure. Two operations may be better, in fact, than a long one-stage operation for some patients with constrictive pericarditis whose condition is poor.

I saw this patient on April 30th, 1945 at the request of Dr. Walsh and at first there was some question in my mind as to whether or not she would be benefited by pericardiectomy. However, inasmuch as she has had a progressively downhill course and since surgery offered the only possible hope of improvement, I agreed to carry out this procedure.

The best results are obtained from operation for constrictive pericarditis in patients with small, quiet hearts. This patient has a large heart and I agree with Dr. Walsh that she does have constrictive pericarditis with co-existing myocarditis. The presence or absence of fluid in the pericardium is of didactic interest only.

My operative note follows: "Operation May 9th, 1945. The patient was placed in a semi-Fowler's position propped up by a number of narrow pillows. The skin incision was made from the lower border of the second cartilage on the right side one half inch from the lower border of the sternum passing downward para-sternally over the third, fourth, and fifth, to the sixth cartilage where the incision was turned lateralwards and downwards to about the mid-axillary line and thence upward into the axilla along the fifth interspace. The muscular structures were divided. All bleeders were clamped and ligated. The third, fourth, and fifth cartilages were divided near the sternum. The internal mammary artery was divided between hemostats and ligated. The fifth cartilage was separated from the sixth cartilage. The pleural cavity was entered through the fifth interspace and the pleura was divided near the sternum corresponding to the incision. Rib spreaders were inserted carefully in such a manner as to avoid pressure upon the cartilages. An excellent exposure of the entire pleural cavity was obtained bringing into complete view the entire right side of the pericardium, the superior vena cave with the phrenic nerves along the lateral border and the pulmonary vessels. The pericardium was obviously distended and numerous small capillary-sized vessels coursing in all directions over the pericardium were seen. Collapse of the lung upon the entrance of the pleural cavity caused no respiratory embarrassment. The pericardium was opened just anterior to the phrenic nerve. About 200 c.c. of straw-colored pericardial fluid was aspirated and a specimen was obtained for guinea pig innoculation. The pericardium was densely adherent to the myocardium by many scattered tufts. There were pockets of fluid varying in size from 5 c.c. to 25 c.c. The pericardium was freed along the anterior border of the phrenic nerve and was then divided along the posterior border of the phrenic nerve leaving this structure intact with a small piece of pericardium attached. The anterior portion of the pericardium was freed from the epicardium as far as the left border of the sternum and the third part of the pericardium was excised all the way up to its attachment to the common pulmonary artery

and the aorta. The pericardium over the right auricle was quite accessible and the dissection was continued around the superior vena cava and downward over the anterior surfaces of the right pulmonary artery and pulmonary veins. Every particle of the constricting pericardium was easily and safely removed. A finger was inserted through the greater transverse sinus and a few rather thin, easily separated adhesions were found. These adhesions were divided digitally and a few additional cubic centimeters of fluid were removed. The left side of the pericardial cavity was explored and numerous short but easily divided adhesions were found. There was an area about the size of a silver quarter on the left side which was not divided. The third, fourth and fifth cartilages were coaptated with O chromic catgut on a cutting needle. The fifth and sixth ribs were approximated with two pericostal sutures of number 2 chromic catgut. Positive pressure was exerted by the anesthetist. A rubber catheter was inserted into the pleural cavity and suction was applied during the remaining part of the closure of the would. The lung expanded promptly. The parietes and the chest wall were closed in layers in the usual manner without drainage. A snug pressure dressing was applied.

Operation: Pericardiectomy, right.

Final diagnosis: Constrictive Pericarditis with Pericardial Effusion."

The operation described above was completed in one hour and eighteen minutes. I do not imply that I am a fast operator but I have made note of this time only to emphasize the absence of the usual technical difficulty resulting from inadequate exposure. The transpleural method of operation permits identification of anatomical structures and clean, easy, sharp dissection.

If this patient continues to have symptoms suggestive of constrictive pericarditis, a left sided approach can be used subsequently and the remaining pericardium removed. However, I do not feel that at this time she has sufficient constriction of the left pericardium to justify additional surgery.

## STILL'S DISEASE

*Case Report No. 29*

Dr. Richard Meyer B. S.—45-1032

B. S., a 21-month old colored female child, was admitted to Children's Hospital on February 9, 1945 with a chief complaint of fever and a rather generalized rash of one half a day in duration. The illness on admission seemed to be related to an attack of pneumonia which had occurred six days previous to her admission. For the pneumonic process an outside physician had prescribed sulfa drug (which one was not known) and the drug had caused a drop in the fever and a good clinical response. However, on the date of admission the fever rose to 103° and the rash appeared. The rash was present on the face, the chest and the knees and was pin-point and erythematous in character. The admitting history was essentially noncontributory since the mother stated that the child had always been well and had suffered none of the usual childhood diseases. Feeding history, growth and development were normal. The delivery of the child had been normal. The physical examination on admission revealed a rather well developed, well nourished colored female child who was not acutely ill in appearance. The positive findings included: (a) slight pallor of the mucous membranes; (b) rash; (c) rapid respiration of 55/min.; (d) râles of a bubbling character in the left lung mainly in the axillary region; (e) a heart rate of 160/min.; (f) liver enlargement of one finger below the costal margin but was not tender; (g) a slightly coated tongue; (h) irritability to active and passive movement; (i) temperature of 105°.

Routine urinalysis on admission revealed nothing but a few white blood cells and epithelial cells. Routine blood count revealed Hgb. of 9 grams, 2.9 million red blood cells, 22,400 white blood cells and a differential of 83% neutrophils, with 72% segmenters and 11% bands and 17% lymphocytes. The tuberculin test (O.T. 0.1 mg.) done at this time was negative. Blood Kahn was negative. A spinal tap was essentially negative.

The initial therapy was directed at a diagnosis of toxic drug rash and resolving pneumonia so that general supportive care was instituted and fluids were forced. The course of the patient during the first hospital week was marked by no clinical improvement. The fever was continuous and ranged from 99.5° to 106°. It was a septic type of temperature. On February 11, 1945 marked posterior cervical lymphadenopathy was noted and a possible diagnosis of measles was advanced. For this reason the patient was put in isolation. This diagnosis failed to materialize.

Because of the continuing high fever and also because a focus of infection could not be found, the patient was put on a course of penicillin for five days beginning on February 14, 1945. Two hundred and forty-five thousand units were given but there was no clinical response, and it was discontinued. The course in the hospital up to the first month was essentially unchanged and in review presented the following features:

(a) Continuous fever of septic type.
(b) Irritability to active and passive movement.
(c) Persistant moderate anemia and leucocytosis.
(d) Posterior cervical lymphadenopathy.
(e) Presence of a rash that desquamated on about the 14th hospital day and led to the advancement of the diagnosis of a possible post-scarlet fever syndrome.

With the above features as a guide, the following laboratory work was performed to determine the focus of infection:

(1) Blood cultures on February 10, 15, 28, March 4, 8, April 22, May 16, 30, and June 1 were all negative.
(2) Spinal tap done on admission and repeated May 8 were both negative.
(3) Febrile antigens were negative in 1/20 titre on February 19, March 5, and May 15.
(4) Urine cultures on seven successive days between March 3 and March 9 revealed no organisms. Repeated urinalysis revealed nothing significant.
(5) Several non-protein nitrogen and blood urea nitrogen studies during the child's stay were well within normal limits.
(6) Repeated blood counts revealed an average of 7–8 grams Hgb. and 2.5–3.0 million red blood cells. This necessitated the administration of 150 c.c. transfusions of whole blood at least once a month.
(7) The white blood cells remained moderately increased and ranged between 9,000 and 17,000 with a predominance of segmented forms.

On March 19, 1945, swelling of the right hand was noticed and at this time the possibility of osteomyelitis was considered. An x-ray of the wrist and hand failed to reveal any changes suggestive of osteomyelitis. An x-ray of this extremity two weeks later was also negative. On March 3, 1945 both ear drums showed bulging and a bilateral myringotomy was performed. At this time the fever came down to normal and remained so for eight days. On the 12th of March the fever again rose and was spiking in character. A second bilateral myringotomy was performed; this time, however, there was no defervescence of the temperature and the fever continued. Repeated x-rays of the mastoid areas showed haziness and a bilateral mastoidectomy was performed on April 26, 1945. Since

that operation, the fever had been a continuous one with daily spikes, returning to normal only once or twice during the next six weeks.

With persistance of fever, the possibility of pyelonephrosis secondary to mastoiditis was advanced. Intravenous pyelographic studies were normal, however, and routine analyses of the urine were negative.

A second course of sulfadiazine and penicillin produced no response. An electrocardiogram at this time was negative except for sinus tachycardia. The child continued to show a septic fever, cervical adenopathy and irritability.

FIG. 4. STILL'S DISEASE
Note fusiform swelling of the proximal digits and swelling of the ankles

On June 17, 1945, definite symmetrical swelling and tenderness of the wrists, knees and ankles appeared. With the joint manifestations showing themselves finally in a full blown fashion, the diagnosis of rheumatoid arthritis was advanced and with the anemia, leucocytosis, early liver enlargement and cervical lymphadenopathy a diagnosis of Still's disease was made. Splenomegaly was not present, however.

The involved joints had a peculiar doughy feel. X-rays of the latter revealed soft tissue swelling but no evidence of bone abnormality. The sedimentation rate at this time was 45 mm./hr. and repeated one month later was 40 mm./hr. A consultation with Dr. B. J. Walsh supported the diagnosis and therapy was directed at the prevention of flexion de-

formities by the use of physio- and hydrotherapy. The child was placed on a diet high in protein, high in carbohydrates and low in fat. Fluids were forced and she was given supplementary vitamins. Fifteen grains of aspirin was given every six hours and this relieved the joint symptoms and lowered the temperature.

## Summary

The case presented the following features: (1) Continuous fever of a septic type; (2) Persistent moderate anemia and moderate leucocytosis; (3) Early rash which was pin-point and erythematous in character and which desquamated in 14 days and failed to reappear; (4) Persistent posterior cervical lymphadenopathy; (5) Early liver enlargement (during first few weeks only); (6) Latent appearance of joint manifestations with definite symmetry of involvement when they appeared. Fusiform swelling of the proximal first phalanx on the left hand; (7) Lack of x-ray evidence of bone deformity or of joint deformity; (8) Presence of definite mastoiditis; (9) Continued irritability of the child to both active and passive movement; (10) Failure of response to chemotherapy; (11) Symptomatic relief to large doses of salicylates; (12) Absence of a definite etiological factor; (13) Elevation of the sedimentation rate.

## Discussion

The diagnosis of Still's disease is often difficult. The typical case of Still's disease may present itself in a myriad of fashions. The presenting symptoms are often vague and may lead to confusion in early diagnosis.

According to Holt and MacIntosh, the predominance of constitutional symptoms such as fever, anemia, lymphadenopathy with irritability for a long period of time without definite joint involvement is the usual rather than the unusual course of the disease. In this case particularly much confusion was bound to exist because the joints showed no definite involvement for almost five months of the hospital stay.

The disease is more common in the age group of 5–7 years but it is not rare at 2 years. The presence of lymphadenopathy is a constant feature in these cases and although it is usually axillary and inguinal it is sometimes confined to the cervical nodes.

The most commonly considered diseases in the differential diagnosis are:

1. Rheumatic fever: This is rare before two years of age. There may be associated chorea and usually there is evidence of carditis. The persistence of joint symptoms is less marked and the joint involvement is migratory in nature.

   Both rheumatoid arthritis and rheumatic fever may show cutaneous

manifestations mainly of the erythematous type, subcutaneous nodules, more common in rheumatic fever and pericarditis. In rheumatoid arthritis there is rarely any pericardial effusion.

2. Periarteritis Nodosa: The clinical picture of this disease may simulate rheumatoid arthritis rather closely. Usually, however, there is nitrogen retention, hypertension and a lack of symmetrical joint involvement. Also there are sometimes palpable nodes along the course of the arteries. A definite diagnosis is usually made on muscle biopsy.
3. Lupus Erythematosis: If the typical skin lesions are present the diagnosis is less difficult. However, if skin manifestations are lacking, malaise, arthritic pains, septic fever, and renal involvement may be the main features.

   Points of differentiation include the presence of a verrucous bacterial endocarditis in some and the presence of leucopenia which is rather constant throughout the course of the disease. Thrombocytopenia and purpura are frequently noted.
4. Other diseases such as septicemia, Brucellosis, typhoid and the dysenteries, tuberculosis, poliomyelitis and osteomyelitis are frequently considered in any individual case.

# LIPOCHONDRODYSTROPHY

*Case Report No. 30*

Dr. Sidney Ross J. D.—37-6776A

J. D., a 5½ year old white male, was admitted to Children's Hospital on June 4, 1941 because of tardy development. According to the patient's mother, he had always appeared somewhat retarded in both growth and mentality. He began to sit up at 16 months, walk at 20 months and talk at 24 months. A partial diminution in hearing was thought to have followed a bout of Scarlet Fever one year prior to entry. There had been no convulsions at any time.

The patient was a full term, 6 pound infant, delivered with forceps, with no history of injury either at birth or post partem. Family history was not contributory.

Physical examination revealed an undersized, underdeveloped child, who appeared grossly retarded both mentally and physically. On standing, he assumed a half crouching position with moderate flexion of the knees and hips.

The head was inordinately long in the AP diameter with prominent frontal bosses and high forehead; the nasion was depressed giving the nose a saddle shaped appearance. There was diffuse clouding of both cornea. Teeth were carious. The neck was short and the patient's head appeared to rest on his shoulders. The trunk was unusually short and there was a noticeable deformity of the thorax characterized by a flaring of the lower costal border with a depression of the xyphoid. A moderate degree of kyphosis was noted. The abdomen was prominent and both the liver and spleen were palpably enlarged. The extremities were unusually short and there was a claw-like deformity of the hands with the phalanges held in constant partial flexion.

Hemogram showed a mild anemia but was otherwise within normal limits. Urinalysis was negative. Blood Kahn was also negative. Total protein was 6 gms. %; cholesterol 166 mg. %; calcium determination was 9.2 mgms. %; phosphorus 3.1 mg. %. Non-protein nitrogen was 40. A glucose tolerance test was performed and was essentially normal. X-ray examination of the skull revealed a very marked enlargement of the calvarium especially in the AP diameter. The sella turcica was definitely elongated measuring 25 mm. This was not associated with any erosion of the clinoid processes. X-ray examination of the long bones showed the centers of ossification to be within normal limits of the chronological age. Both acetabulae and glenoid fossae were shallow. There was a

definite kyphosis of the spine in the dorsal and upper lumbar region with narrowing of the intervertebral spaces of the last three dorsal segments. There was some wedging of the body of the third lumbar vertebra.

It was thought that these findings associated with the clinical appearance of the patient pointed rather definitely to a diagnosis of lipochondrodystrophy. Since that time the patient has been followed at periodic intervals in the out patient department by Dr. John Washington.

FIG. 5. DYSOSTOSIS MULTIPLEX (HURLER'S SYNDROME)
Note kyphosis and typical flexion deformity of the knees and the peculiar shaped skull and hands

## DISCUSSION

Dr. John Washington: This child definitely suffers from the disease first described by Hurler in 1919. The deformities characteristic of the disease vary greatly in degree and this patient is less crippled than the average. The diagnostic features are enlargement and misshaping of the head, shortening of the trunk with kyphosis behind and a protruding abdomen in front, enlargement of the liver and spleen, limitation of motion

of joints usually most evident in the fingers, and clouding of the cornea. Retardation in growth becomes noticeable about the fourth year. Severe dwarfism occurs in some cases while in others the height is only mildly affected. Mental retardation usually is present. Nasal stridor is often a conspicuous feature.

X-ray findings vary with the severity of the clinical involvement. In the very crippled cases the shaft ends are distorted and the shafts, particularly the ribs, seem broader than normal and all the bones laid down in cartilage when compared with bones of normal children of the same age appear under-developed and clumsily moulded. The scapulae especially appear small with blunt acromion and coracoid processes. The vertebral body at the site of kyphosis (usually the first lumbar vertebra) becomes wedge shaped due to a failure of anterior growth of the upper table, leaving the lower table protruding as a lip to form the apex of the wedge. X-rays of the enlarged head often show a peculiarly elongated sella turcica. In the less severely crippled cases, as the one here reported, x-ray findings are much less dramatic, the striking findings being the wedge shaped vertebra, the elongated sella turcica and the rather wide appearing ribs. Pathologic studies of the epiphyses in one case showed delayed and irregular growth of cartilage cells.

The clouding of the cornea is an expression of a feature of this disease which seems entirely separate from the skeletal abnormalities. On pathological examination it is found that in the cells of the brain there are degenerative changes with the deposition of granules of some lipid substance. Similar findings occur in amaurotic idiocy and Nieman-Pick's disease. The clouding of the cornea is apparently due to deposition of this substance there. Microscopic sections show similar depositions throughout the reticulo-endothelium system. The deposits outside the brain are not so heavy as in Nieman-Pick's disease, however.

Any attempt to classify this bizarre disease is difficult. Roentgenologists unhesitatingly make the diagnosis of chondrodystrophy in the severe cases which show the irregular shaft ends characteristic of classical chondrodystrophy and of Morquio's disease. In the less severely affected cases the generally accepted criteria for an x-ray diagnosis of chondrodystrophy are lacking. However, it would appear that the criteria should be broadened to include the features seen in these less severely affected cases. Some cases have been reported as examples of Morquio's disease. The skeletal deformities with the exception of the enlarged head would fit this diagnosis but it is evident that the additional features demand a separate classification. Hurler's disease, dysostosis multiplex, gargoylism, and lipochondrodystrophy are terms in use at the present time. The latter term is descriptive of the pathologic lesions.

The prognosis is hopeless as far as cure is concerned. No indication for endocrine therapy exists in theory or from the results of metabolic and pathologic studies. Thyroid extract is reported to have improved the mental state of one or two cases but in general it causes no change. Because the disease has a strong tendency to repeat itself in sisters and brothers, parents should be warned of the risk involved in further propagation. As the occurrence of the disease apparently depends upon the chance union of two individuals, who, unknown to themselves, each carry a rather rare recessive gene, it is probable that children of either parent by some other mate would be normal.

## CLINICO-PATHOLOGICAL CONFERENCE

Directed by Dr. E. Clarence Rice

Assisted by Dr. John E. Cassidy

Held every Tuesday afternoon at 3:00 p.m.

*Case Report No. 31*

Dr. John E. Cassidy D. G.—43-9402

D. G., a three-year old white male, was admitted to the hospital on December 3, 1943 with the chief complaint of limping. Two days prior to this he had fallen down a flight of four steps and since than was noted to have a slight limp and to have some difficulty in climbing stairs. X-ray studies of the pelvis and femora done on the day of the fall failed to reveal any evidence of fracture. After the fall the mother noted that there was quite a marked decrease in the child's activity. Whereas usually he was constantly running about and playing he now was content to sit in a chair or recline on a couch and resented very much being disturbed. His past history was negative except for measles six months previous. Family history revealed one sibling who was living and well. The parents were both living and apparently in good health.

Physical examination showed a well developed and nourished white boy who did not appear acutely ill. His temperature was 99.8° (R), pulse 90, and respirations 20. The physical examination was essentially negative except for slight nuchal rigidity.

Laboratory work revealed the urine to be normal; hemoglobin of 13 grams and red-cell count of 4,110,000; white-blood count 8,900 with 37% neutrophils, 52% lymphocytes and 10% monocytes. Spinal fluid examination revealed 205 cells per cu. mm., 36% polys. and 64% lymphocytes, 200 mgm. of protein per 100 c.c., and a reduction in the sugar content.

Hospital Course: During the first twenty four hours the patient was alert and cooperative and ate well. During the second day irritability was noted. A repeated spinal fluid examination revealed essentially the same findings as the initial one. Irritability increased and periods of marked listlessness appeared. Repeated urinalyses were negative and the blood picture remained unchanged. A low grade fever developed, the temperature ranging between 100° and 102°.

On the morning of the seventh day a generalized convulsion occurred lasting twenty minutes. The next day convulsions again appeared and were continued intermittently until death the following day. Terminally his temperature rose to 107°. X-ray examination of the chest two days

prior to death was reported negative except for increased fibrosis at the roots of the lungs. A tuberculin test, 0.1 mgm. of old tuberculin, was negative.

Necropsy Findings: The body was that of a well developed and nourished white male child. Opalescent cisternal fluid was obtained which showed essentially the same changes as the spinal fluid specimens obtained before death. A Levinson test done on this fluid was positive. Cultures from all of the specimens of cerebro-spinal fluid yielded acid fast bacilli.

The important findings were confined to the central nervous system and the thorax.

There were many small pearly gray tubercles scattered over the surface of the brain, being more prominent along the course of the vessels. At the base of the brain there was a small amount of greenish gelatinous exudate typical of that seen in tuberculous meningitis. In the posterior portion of the lower lobe of the left lung there was a small, firm, yellowish fibro-caseous lesion measuring 1 cm. in diameter. The regional lymph nodes showed small areas of caseation necrosis. The mediastinal lymph nodes on the left side showed similar changes.

Microscopic examination of these tissues showed changes typical of tuberculosis. The pia arachnoid showed small areas of caseation and definite tubercles with numerous giant cells. Several of the blood vessels showed adventitial inflammation. The lesions in the lung and adjacent lymph nodes also showed microscopic changes typical of tuberculosis.

## Discussion

This case is presented to illustrate how easily one can be misled. Attention was at first focused on the hip in this child because of the fall but it soon became evident that the trauma which had occurred was most likely the result of some other pathology which was present. The onset can justifiably be called abrupt and the course rapid, death occurring two weeks after the onset of symptoms. There was no history of any contact with tuberculosis but after the diagnosis was confirmed at autopsy investigation of the family showed that the father had an active infection.

X-ray examination of the lungs failed to pick up the primary lesion and the tuberculin test was negative. Negative x-ray findings and tuberculin tests are noted occasionally in the miliary form of tuberculosis. The infection evidently was not of sufficient duration for this child to have developed a sensitivity to tuberculin.

Most often tuberculous meningitis is secondary to or a part of a generalized miliary tuberculosis. In this case there was no miliary involvement other than that of the pia arachnoid. The pulmonary lesion was the type that is commonly regarded as a primary complex.

The characteristic course of patients with tuberculous meningitis is attended by irritative symptoms initially with listlessness and stupor later becoming more pronounced. At first they may be roused but finally the stupor becomes so profound that it is impossible to arouse them. Involuntary movements of the arms are frequently noted. The course in this case was somewhat shorter than usual and characterized by irritability and convulsions. The difficulties that may be encountered in making an early diagnosis of this condition are well brought out by this patient.

The spinal fluid findings resemble those seen in lymphocytic choriomeningitis, the various types of encephalitis and meningoencephalitis and poliomyelitis. The organisms were not demonstrated by direct smears of the cerebrospinal fluid but were cultured from three different specimens after death.

During the year 1944 at Children's Hospital there were nineteen cases of tuberculous meningitis, all of whom died.

In this series, fourteen were colored, five white. Fourteen cases were under two years old, the other five under six years of age. There was no elicitable history of contact in eight cases or thirty eight percent. Spinal fluid examination uniformly revealed a pleocytosis ranging from forty to five hundred cells with lymphocytes predominating; protein content was elevated, with sugar and chlorides depressed in the majority but not in all instances. The Levinson test was performed in nine cases of which four were negative, and five positive. Interestingly enough, the tuberculin test was negative in five of the nineteen cases, an observation which is in keeping with the well known fact that a tuberculin test may be negative in fulminating tuberculous involvement.

Treatment of tuberculous meningitis with diasone, promin or promizole was not attempted in any of our cases since recent reports in the literature warrant the conclusion that the efficacy of these drugs in the treatment of tuberculous meningitis is negligible.

# CLINICAL PROCEEDINGS OF THE CHILDREN'S HOSPITAL

## Washington, D. C.



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Occasionally, the remarks and observations of guest speakers are included in this bulletin when thought to have particular interest. The proximity of the Children's Hospital to the Medical Centers of the Army, Navy and United States Public Health Service affords us the opportunity to invite many distinguished physicians to our conferences.

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# BACKGROUND

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